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EXENCEPHALIC (INIENCEPHALIC?) MONSTER WITH BILATERAL HARELIP AND CLEFT PALATE.[1]

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THE monster which I wish to present this evening is a seven months' white female fœtus which was stillborn, being the first child of an extremely neurasthenic woman of seventeen and a half years, whose family history I cannot obtain, but whose pregnancy up to the time of the miscarriage was uneventful. The father is about twenty-five years of age, healthy and robust, but having a strong family history of tuberculosis, his father having died of phthisis, and his mother is now suffering with the same disease.

About one week before the miscarriage the mother fell directly forward upon her abdomen, followed by severe lumbar pain and uterine hemorrhage, but continued to feel fœtal movements up to within two days of the birth. Hydramnios was not present, as has been frequently reported in cases of this malformation, and labor was not long or severe.

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[1] Read at the meeting of the Philadelphia Pathological Society, held February 13, 1902.

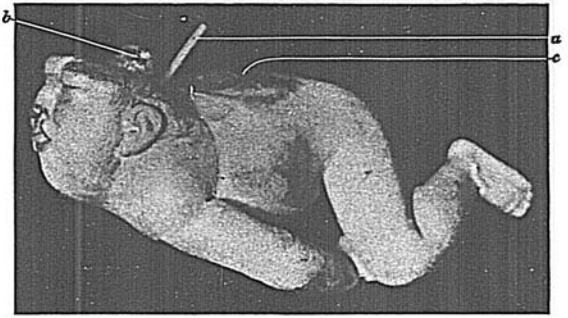
Following the classification of Isidore Geoffroy Saint Hiliaire, whose work upon teratology has practically been accepted and followed by all writers upon this subject, this monster belongs in the general class of exencephalus, which is described as "characterized by a malformed brain situated at least in part without the cranial cavity, the bony walls of which are themselves imperfect." This general class is further subdivided into (1) the notencephalus, in which the cranial contents are in large part without the skull, resting upon the back of the neck, the tumor protruding through a fissure or aperture in the occipital bone. This deformity is usually associated with hydrocephalus of greater or less degree; (2) proencephalus, in which the same deformity as just described exists upon the anterior portion of the skull; (3) podencephalus, and (4) hyperencephalus, in these the cranial deformity is upon the vertex and the tumor protrudes from above. The podencephalus differing from the hyperencephalus only in degree; (5) iniencephalus, in this subclass the deformity of the skull is the same as in notencephalus, with the addition of a spinal cleft of varying degree; in some cases the failure of union extends as far downward as the coccyx, leaving the spinal canal open throughout its entire length. In this class lordosis and compensatory kyphosis are common—the head is tilted strongly backward, causing the face to look almost directly upward, which, with the faulty development of the vault of the cranium and apparent, rather than real, protrusion of the eyes, has suggested the term "frog-faced" monster, although this term is not exclusively applied to the iniencephalic subclass.

In 1896 Dr. H. F. Lewis presented a specimen of this monstrosity to the Chicago Pathological Society, and at the same time collected all the recorded cases which, with two other specimens, he described at the same time and which had not previously been recorded, brought the total number to twenty-five. I have been able since that time to find the report of only four other cases, although the search through the records has been cursory rather than exhaustive.

No mention could be found in any of the recorded cases of an associated harelip or cleft palate (unless the case described by Dr. Wood could be looked upon as an attempt at harelip formation), with the exception of a single sketch in Ahlfeld's *Atlas* which quite closely resembles the photograph of the present specimen.

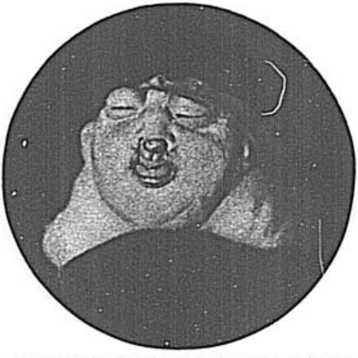
The present specimen is an under-developed female fœtus; skin and visible mucous membranes normal. The neck is entirely absent, the malformed head being placed directly upon the upper portion of the thorax in such a position as to turn the face directly upward, so that the plane of the face is at a right angle to the plane of the body, and to cause the deformed or under-developed occipital bone to rest directly back upon the anteriorly curved cervical vertebræ (Figs. 1 and 3).

FIG. 1.



The general appearance of the monster. The match-stick (*a*) has been passed into the opening in the malformed brain cavity; (*b*) the remains of the brain, meninges, and fringes of skin rolled up as a ball; (*c*) area over which there was failure of development of true skin. The constriction about the upper portion of the thorax and the arm was produced by a tight string used to suspend the fœtus in the preserving fluid.

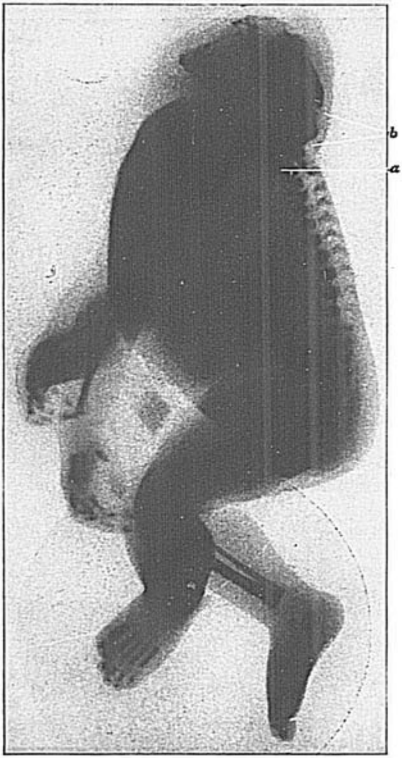
FIG. 2.



Photograph of monster, showing the characteristic formation of the face, and in addition the bilateral harelip, with the greatly protruding intermaxillary bone. The two white spots seen just below the centre of the tongue were caused by moisture on the lip at the time the photograph was taken.

The entire upper portion of the skull above an imaginary plane beginning on a level with the superciliary ridges and passing backward at a right angle to the plane of the face is entirely absent and replaced by a solid bony plate covered by hairy skin. At the junction of the

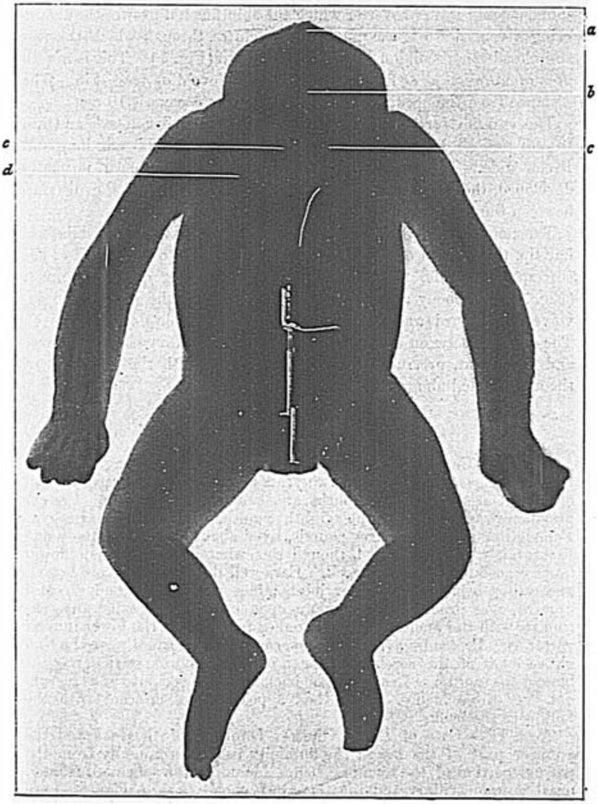
FIG. 3.



"X-ray" photograph, showing (a) the characteristic cervical lordosis. The remains of the brain and meninges appear as the faint shadows surrounding the optical bones (b).

occipital bones and spinal column is an opening into the skull large enough to admit the end of one's little finger (in the photograph is shown a match-stick passed into the opening), from the margins of this open-

FIG. 4.



"X-ray" photograph, showing (*a*) the protruding intermaxillary bone; (*b*) failure of union of hard palate; (*cc*) abnormal position of malformed and separated optical bones; (*d*) scapula and scoliosis.

ing are extending the brain membranes and remains of the brain, together with fringes of skin (in the photograph these are seen rolled up as a ball).

Extending over the back from the opening into the skull to the lower lumbar vertebræ and laterally about 2.5 cm. to each side of the spinal column is an area over which the skin has not grown, the tissues being absent down to the muscular structures, through which the ribs and closed spinal column can be plainly seen (Fig. 1). This is not an area of abrasion as at first appears, but one devoid of true skin. The distance from the opening into the skull to the coccyx is 10 cm.

The ears, so far as can be seen, are normally developed, and are behind and slightly below the shoulders. They are 2.5 cm. long and 1 cm. wide. The eyes are normally located and appear normally developed, the palpebral fissure measuring 1.5 cm. and the distance between the inner canthi is 1.5 cm.

The mouth, which is 2 cm. wide, is the seat of a well-marked bilateral harelip, the cleft on each side extending well up into the nostril of the corresponding side, the intermaxillary bone being quite prominent (Fig. 2). The bony cleft extends backward into the mouth and involves both hard and soft palate to a considerable degree (Fig. 4). The distance between the alæ nasi is 1.5 cm. The tongue is quite large and prominent, protruding from the mouth. With the exception of the above-noted abnormalities the fœtus is normal.

Synopsis of Collected Cases.

Case I.—(Case of Dr. Albert C. Bowerman.) Patient, aged twenty-four years, gave birth, about two years ago, to an apparently normal child, but it died in ten months of some cerebral trouble. Present pregnancy advanced to the eighth month. Hydramnios present. Female fœtus, weighed three pounds, lived about one hour. Body and limbs fairly well developed, though somewhat disproportionate, lower limbs especially long. No neck. Ears well developed and are behind and rather below the shoulder joints; face well formed and directed upward, eyes prominent. Cranium open in occipital region and continuous with the unclosed spinal canal as far down as the lower lumbar vertebræ. Protruding from the upper end of this canal were the congested lobes of the cerebellum much enlarged and exposed across the base of the sacrum. Length of the back from the occiput to the sacrum not over three inches. Child had appearance of crescent with the hollow at the back.

Case II.—(Case of Dr. B. Onuf.) Duplicity of the face and the anterior part of the head; the duplicity begins proximally from the petrous portion of the temporal bones, including the sphenoid, frontal, nasal, supramaxillary and inframaxillary bones. Two pharynges are present which communicate with each other. Craniorrhachischisis involves the entire vertebral column.

There is the interposition of a peculiar os sacrum-like vertebral formation, and two longitudinal bones (each divided into anterior and posterior portion) between the occipital and temporal bones of each side. In the proximo-distal direction these intercalary formations form the transition between the bones of the skull and the vertebra. There is also absence of the bony sacrum.

CASE III.—(Case of Dr. W. Atkinson Wood.) Mother, aged twenty-three years; father, aged twenty-six years. Female fœtus born dead, but the tissues appeared healthy and fresh. Entire scalp from the superciliary ridge to the position of the occipital protuberance and from one mastoid region to the other was a mass of dry gangrene studded with pear-shaped fibrous outgrowths. Both eyes protruded markedly, and the mouth in the mid-line was quite absent, being represented by two lateral slits into which a probe passed easily for half an inch. Both ears were well developed. At the third or fourth lumbar vertebra was a myelocele. The lower extremities were fully flexed at the hips, so that the feet which were in the position of talipes spasticus, or dorsalis, were fitted and pressed accurately under the position of the chin.

The vertex was removed on a level with the superciliary ridge, and the bones were so fused together as to appear as one bony mass. The petrous portion of the temporal bone and mastoid bones were fused into one mass; on the inner side of this mass were two cavities containing brain matter, each about the size of a bean. In the substance of these bony masses the ossicles were well developed. The sphenoidal cells were well developed, and the nasal chambers connected directly with the anterior fossa of the skull.

The vertebral column had no direct communication with the skull, being kept in apposition only by skin and connective tissue. The spinal canal opened directly at the depression on the posterior part of the skull, the spinal cord being thus exposed to the air.

The pharynx and larynx were well developed. Tongue and tonsils were contained in a membranous sac on the anterior surface of the spinal vertebræ. The internal carotid was doubtfully developed (which the author ascribed as the probable cause of the cranial deformity).

The lower and upper jaws were ankylosed together; the buccal cavity was divided into two lateral halves. The alveolar processes of the upper jaw were fused with the hard palate, leaving a slit-like opening between.

CASE IV.—(Case of Dr. Barton C. Hirst.) The doctor presented a photograph of the monster, with no description.

The specimen shown this evening will be presented to the Wistar Institute of the University of Pennsylvania.

I am indebted to Dr. Clifford B. Farr for the translations of the foreign references, and to Dr. Thomas S. Stewart, who furnished the "X-ray" negatives.

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A CASE OF HYPERNEPHROMA OF THE KIDNEY COMPLICATING PREGNANCY.

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and

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of Philadelphia.

The following case of tumor of the kidney appears to deserve mention both because of the rarity of the tumor and the peculiar manner in which it manifested itself.

Mrs. T., a negress, aged thirty years, was admitted as an emergency case to the Medico-Chirurgical Maternity Hospital, on November 3, 1900. Examination of the patient on admission showed her to be about six months pregnant. She complained of pain over the uterus, and an internal examination showed evidence of a threatened miscarriage. More interesting than the pregnancy was the presence of a tumor which could be felt at the lower border of the liver. Dulness over this tumor extended into the right flank.

We were unable to elicit any previous history throwing light upon its origin. The patient was anæmic, but had not the characteristic cachexia of malignancy. The urinalysis was negative, the temperature elevated. Five days after admission she spontaneously miscarried, giving birth to a six months' fœtus. After delivery the tumor stood out more prominently and appeared to be rapidly increasing in size.

A tentative diagnosis of either abscess of the liver or a large growth of the kidney was made. The tumor had not the feel of a cystic kidney, nor were the clinical symptoms suggestive of liver abscess. She still continued to have fever and was rapidly growing worse, and on November 17th was etherized and an exploratory incision made. The tumor occupied more of the region of the liver, and upon deep palpation fluctuation was noticed.

An incision was made over the most prominent portion of the growth through to the peritoneal cavity. What was then found was supposed to be a retroperitoneal abscess, so gauze was packed around the wound to shut off the general peritoneal cavity, and an incision was made into this bulging mass. A large quantity of thin, dark, very offensive fluid escaped. It had the odor of urine. As the patient was now in an extremely critical condition, a large drainage-tube was inserted. The abscess was drained, and the next day the patient seemed a trifle improved, but she commenced to lose ground again, and died December 17th, a month and a half after her admission to the hospital.

Autopsy Notes. December 19, 1900. Body of a much emaciated young colored woman of middle stature, about thirty years of age. The face bears an expression of suffering. Rigor mortis is absent. Inspection of the body shows an open wound in the right hypochondrium, large enough to admit a finger, and communicating with some part of the intestine, forming a fecal fistula. The edges of the wound are indurated, the surface necrotic and foul. In addition to this intestinal fistula, there is a sinus which descends deeply toward the right